

Case Report

Presentation of acute central retinal vein occlusion in scleroderma

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Abstract

Central retinal vein occlusion (CRVO) is a rare complication of scleroderma. Here we report a case of a 30-year-old man who was diagnosed to have scleroderma in the rheumatology and dermatology clinic. During treatment with systemic steroids and immunosuppressive therapy the patient developed a sudden decrease of vision in the right eye and was diagnosed to have right CRVO with macular edema on fundus examination. After three consecutive Intravitreal bevacizumab (IVB) injections for macular edema, best-corrected visual acuity (BCVA) improved from 20/80 to 20/25. All ocular and systemic causes of CRVO other than scleroderma were excluded in our patient by thorough clinical examination and investigations, suggesting that scleroderma was the most possible etiology in his condition.

Keywords: Central Retinal Vein Occlusion, Scleroderma, Systemic sclerosis, Eye manifestations, Intravitreal bevacizumab (IVB), Macular edema

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Introduction

Scleroderma or systemic sclerosis (SSc) is a generalized chronic connective tissue disorder of unknown etiology.¹ One of the critical abnormalities in SSc is a diffuse fibroproliferative vasculopathy, most commonly involving the microvasculature system,² which includes perivasculitis and fibrinoid degeneration of blood vessels.¹ Scleroderma usually appears in patients between 35 and 55 years, women being 2–3 times more frequently affected than men.¹

Scleroderma patients commonly report eye problems, with dry eyes being a typical complaint. Conjunctiva and eyelid abnormalities are also often seen, including injection, telangiectatic vessels and stiff or tight eyelids. Diffuse iris atrophies and lens opacities are also experienced, although the use of corticosteroids as part of treatment is likely to

be the cause of the latter of these.¹ However, involvement of the posterior segment is often subclinical and visual loss as a direct result of the disease is rare.

Here we report a case of Central retinal vein occlusion (CRVO) with decreased vision, which is a rare complication of scleroderma. Patient was treated with Intravitreal bevacizumab (IVB) injections for macular edema and responded well with vision improvement. To our knowledge, CRVO with scleroderma was reported only twice in the literature.^{3,4} We herein report a third case with that very rare complication.

Case report

A 30-year-old man was diagnosed as scleroderma in the rheumatology and dermatology clinic three years ago. He

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presented with features of thickening, tightening and induration of the facial skin. There were associated sclerodactyly, periungual telangiectasia, digital pitting, calcinosis in the right fourth finger and positive Raynaud's phenomena. History was negative for any medical problems other than scleroderma.

All investigations at presentation which include thyroid functions, complete blood count (CBC), prothrombin time (PT), partial thromboplastin time (PTT), random blood sugar (RBS), renal function tests (RFTs), liver function tests (LFTs), rheumatoid factor, HLA-B51, HLA-27, anti-double-stranded DNA, anti-scleroderma antibody, anti-JO-1, anti-ribonucleoprotein, anti-Sjogren's antibody SSA, Anti-Sjogren's antibody SSB, C3, C4 complement levels, centromere protein antibody, Smith antibody, anti liver/kidney microsomal antibody (ANA) speckled ((1:320)), erythrocyte sedimentation rate (ESR) 67 mm/h normal (0–20) mm/h and C-reactive protein (CRP) 0.4 mg/dl normal (0–0.3) mg/dl.

A treatment in the form of oral prednisolone and mycophenolate mofetil (Cellcept) was started since that time.

As the patient was on routine follow up in the ophthalmology clinic for dry eye, an incidental finding of tortuous retinal vessels in both eyes on fundus examination was documented.

Two years after the patient was diagnosed to have scleroderma, he presented to our clinic with a sudden decrease of vision in the right eye of one-day duration. On examination his Blood pressure was 125/75 mmHg, and best-corrected visual acuity (BCVAs) were 20/80 and 20/20 in the right and left eyes, respectively. Both pupils found to be normal with no relative afferent pupillary defect (RAPD). Anterior segment was within normal for both eyes. Intraocular pressure was normal in both eyes. The fundus of the right eye showed tortuous vessels with multiple retinal hemorrhages, cotton wool spots and optic nerve swelling (Fig. 1). Fundus fluorescein angiography (FFA) showed capillary nonperfusion of less than 10 disk diameters. Macular edema and neurosensory detachment were also detected in the right eye by optical coherence tomography (OCT) (Fig. 2). Fundus examination of the left eye showed mild tortuous vessels (Fig. 3).



Figure 1. Tortuous vessels with multiple retinal hemorrhages, cotton wool spots and optic nerve swelling in the fundus of the right eye.

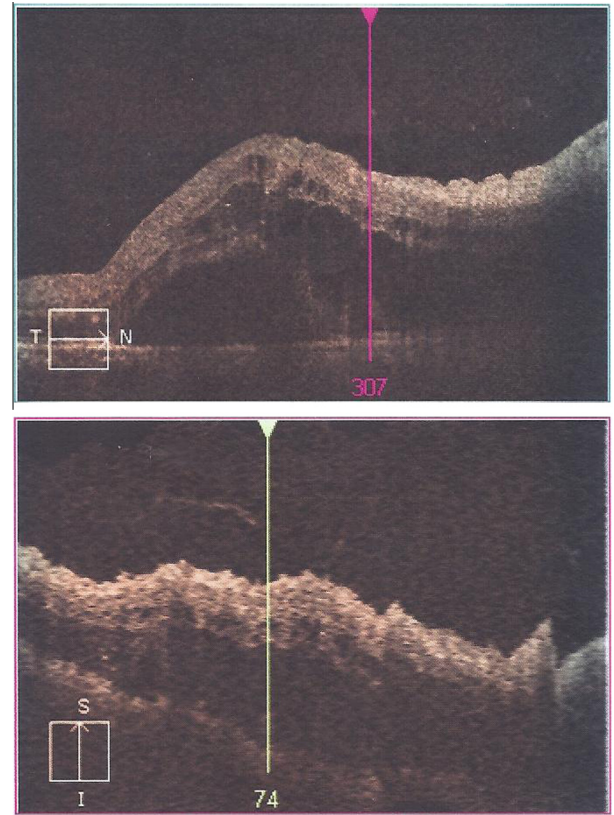


Figure 2. OCT of the right macula on presentation showing macular edema with neurosensory detachment.



Figure 3. Mild tortuous retinal vessels of the left eye.

Based on the previous findings, a diagnosis of non-ischemic CRVO in the right eye was made, in which the patient received three consecutive IVB injections for the macular edema on monthly basis.

After six-months of follow up, his right BCVA was improved to 20/25 and the macular edema was resolved (Fig. 4). Retinal hemorrhages, cotton wool spots and optic nerve swelling were resolved leaving only a track of some sclerosed blood vessels with no neovascularization in the retina, iris or the iridocorneal angle. Intraocular pressure was

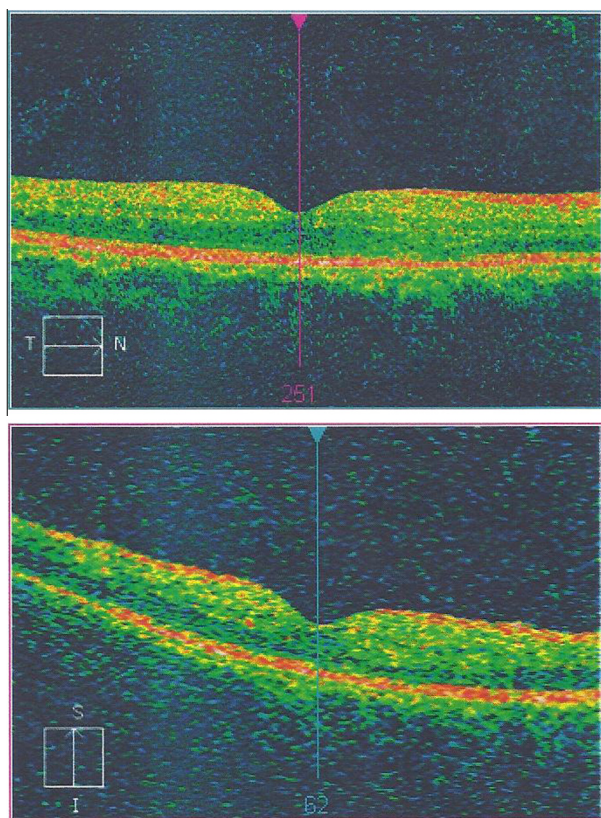


Figure 4. OCT of the right macula with normal thickness 6 month after treatment with three consecutive IVB injections.

within normal range. All causes of CRVO were excluded such as hypertension, diabetes, glaucoma, blood dyscrasias, clotting disorders, sickle cell disease and retinal vascular diseases.

Discussion

Scleroderma is a systemic connective tissue disease of unknown etiology. It is characterized by fibrous and degenerative changes in the skin and viscera.¹ The most characteristic feature of scleroderma is the increased collagen content in the skin and visceral organs. The second characteristic feature is vascular disease, including Raynaud's phenomenon and diffuse microvasculopathy.⁵

The American College of Rheumatology provides diagnostic criteria for SSc. Major criteria include anterior scleroderma (thickening of the skin) affecting the arms, face, and/or neck; and minor criteria involve digital pitting scars or loss of the volar pads of the finger tips, bibasal pulmonary fibrosis and sclerodactyly (sclerosis affecting the fingers or toes). SSc is diagnosed when a patient has one major or two minor criteria.⁶

Our patient fulfilled these criteria as he was having tightening and induration of the skin of his face, sclerodactyly, periungual telangiectasia, digital pitting, calcinosis and positive Raynaud's phenomena.

The ocular manifestations of SSc are characterized by marked heterogeneity. Eyelid skin abnormalities and keratoconjunctivitis sicca (KCS) are the most common SSc-related ocular findings. In the posterior segment of the eye, retinal

microvascular abnormalities indistinguishable from those related to systemic hypertension are the most common findings, and they seem to be associated with a severe capillaroscopic pattern.⁷ In our case the patient was on follow up for dry eye and discovered incidentally to have tortuous retinal vessels. Tortuous vessels documented before the presentation of CRVO indicates an abnormal retinal microvasculature.

A study of the retinal vasculature of scleroderma patients using fluorescein angiography demonstrated abnormalities of perfusion, which affected the choriocapillaries and small choroidal arterioles. Most of these patients were normotensive. These observations suggest that end-organ disease due to retinal vessel involvement with scleroderma vascular change does occur, even though uncommonly.⁸

CRVO affects 0.8 in 1000 people, and presents with variable degrees of visual loss.⁹ Funduscopic exam typically shows numerous retinal hemorrhages, dilated and tortuous retinal veins, cotton-wool spots and the presence of macular and optic disk edema.¹⁰ CRVO in our case was a non-ischemic type and showed favorable prognosis after treatment with three consecutive IVB injections. It ended with no severe complications such as neovascular glaucoma.

Ushiyama et al. studied the retinal findings in 29 patients with SSc and reported a higher incidence of retinal changes associated with vascular damage in SSc patients than in controls ($p = 0.01$).¹¹

CRVO in scleroderma is a rare complication, which was found in our case and was reported only twice in the literature by Saari et al. and Littlejohn et al. in 1981.^{3,4} In our case the patient had no other diseases, which could be the cause of the CRVO, in contrast to the cases reported by Saari et al. and Littlejohn et al. in which the patients suffered from other systemic problems such as secondary polycythemia, cardiovascular insufficiency and pulmonary fibrosis.^{3,4}

Endothelial cell dysfunction, which affects microvasculature system, could be the cause of CRVO in our case. Thorough history and examination of the skin and joint are essential in a patient with CRVO.

In conclusion, continuous follow up is essential for patients with scleroderma presenting with abnormal retinal vasculature.

Conflict of interest

The authors declared that there is no conflict of interest.

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